THE CEREBRAL LESIONS IN A CASE OF COMPLETE ACQUIRED COLOUR-BLINDNESS

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(*From the Laboratory of the Royal College of Physicians, Edinburgh*)

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WE publish this record of a case where colour vision was completely lost, while some form sense, achromatic vision, persisted, believing it to be the first complete case of the kind that has been submitted to *post-mortem* examination. As far as we are

aware only one comparable case of acquired colour-blindness has been examined *post-mortem*, and recorded. We refer to that by Verrey (*Arch. d'ophthal.*, xvii., 1888, p. 289). His case differed from ours in that it was one-sided (only hemiachromatopsia), and showed a more extensive lesion of the cerebrum.

Our patient, aged sixty-two, the managing director of a large and complicated commercial undertaking, was a man of great mental capacity; he had enjoyed good health, and had previous to his last illness suffered from no malady which could in any way affect his sight. His colour sense had always been good. He always took a great interest in the management of his garden, and was able to appreciate the most delicate colours in his flowers.

In 1896 hard work and excessive worry in the management of his business affairs seemed to have reduced his strength, he became anæmic. Early in 1897 symptoms of dyspepsia were added to those of anæmia. No definite diagnosis of his case was made until November of that year, when a carcinoma of the stomach was found. The symptoms of debility and dyspepsia increased, and at the time of his death, April 1898, he was very emaciated. Thus far the history of the case is a fairly typical one of carcinoma of the stomach.

An affection of the sight was first noticed on November 24th, 1897. On the previous day his eyesight had been as usual, but on the morning of the 24th he found himself unable to read his letters, and when out walking later in the day it was found that he was unable to recognise vivid colours in some children's dresses. His eyes were tested that afternoon (not by the authors), and found to be completely insensible to colour.

On December 23rd, 1897, he was examined by Dr Dunlop, and found to be completely colour-blind. All coloured objects appeared grey. He was unable to recognise the colours of brightly-coloured cushions. He could not see mustard on his dinner plate. He could not tell by sight whether a fire in the grate of his room was glowing or not. His distant form sense as tested by Snellen's types seemed good. He had difficulty in reading continuous print even with the help of his usual presbyopic glasses.

On January 26th, 1898, Dr Mackay saw him, along with Dr Dunlop, and noted his condition as follows :—

External aspect of eyes natural. No obvious signs of blindness. Patient recognised faces, and walked about freely.

Pupils medium size, equal, reacting to light and on convergence.

Ext. musc. — Movements good.

Oph. Exam.—Media clear. Optic discs slightly senile and anæmic, but not atrophic. Vessels well proportioned. Fundus generally healthy. Refraction emmetropic.

$V = \frac{6}{12}$ Sn. with each eye separately, and attempts at $\frac{6}{9}$ Sn. $\bar{C}$ + 4 D sph. = J. 1 with each eye.

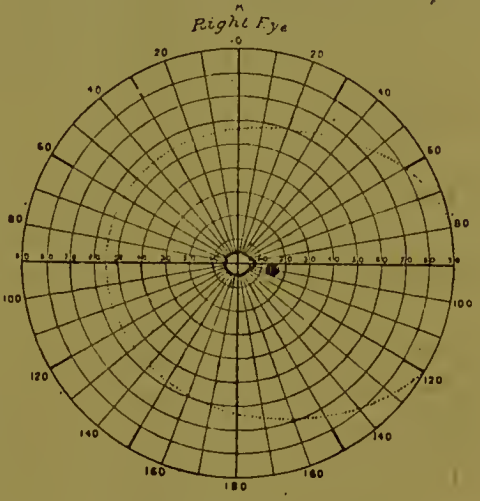

FIG. 1.—Field of right eye for 1 cm. sq. of white paper.

Fields of Vision.— Fields for hand movements *apparently good*; for counting fingers eccentrically curtailed; for a 1 cm. square of white paper restricted in the right eye to an area not exceeding 5° inwards, upwards, and downwards, and to 7° outwards (*i.e.* to the right) from the fixation point; in the left eye to an area which only exceeded 10° at the inner side extending there (*i.e* to the right) to 15° in the horizontal plane.

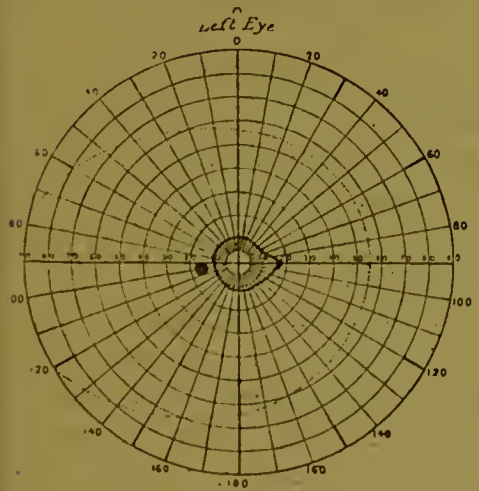

FIG. 2.—Field of left eye for 1 cm. sq. of white paper.

The fields were taken with a Priestly Smith's perimeter. The size of the fields may have been influenced to some extent by fatigue, the left eye having been examined first. If we regard the case as one of double homonymous hemianopsia, the left half fields of both eyes were more affected than the right.

Eccentric Form Sense, tested by recognition of a continuous black band (2 cm. × 8 cm.) on a white surface contrasted with an interrupted black and white band (2 cm. of black squares separated by 2 cm. white squares)[1] was found to be limited to the fields for white, and his perception was rather better below than above the horizontal plane.

Colour Sense.—Completely suppressed for all colours all over the fields. Every colour appeared grey—blue described as glass.

Light Sense.—Not specially investigated for want of proper means at the patient's house.

Intellect.—Clear. No word blindness. Memory of colours seemed good. Quite conscious of his lost sense.

Diagnosis.—Double homonymous hemianopsia, complete for colour sense, incomplete for form sense about the point of fixation, probably due to symmetrical cortical lesions in or near the calcarine fissures, with perhaps slightly more extensive destruction of the right cerebral visual area than of the left.

As to the actual character of the lesions, we considered but put aside the probability of an apoplectic effusion or cerebral tumour, and concluded in favour of vascular occlusion, not without some hope that the defect might be to some extent dependent on his anæmic condition.

During the two months following this examination he acquired greater facility in reading, but there was no restoration of his colour sense.

On April 17th he suddenly developed a right-sided hemiplegia, and on April 22nd died.

The *post-mortem* examination was made twenty-four hours after death. The brain was hardened in ten per cent. formaline solution. At the *post-mortem* examination, the diagnosis of carcinoma of the stomach was confirmed.

The brain was examined both for some lesion to explain the hemiplegia, and for lesions to explain the double hemianopsia. No lesion explaining the hemiplegia was found. The regions of the brain occupied by the motor areas and motor tracts appeared on examination to be healthy. The optic nerves were examined, and found to be healthy. Both occipital lobes, however, the lobes which are undoubtedly connected with

[1] See *Brit. Med. Jour.*, Nov. 10, 1888, p. 1036.

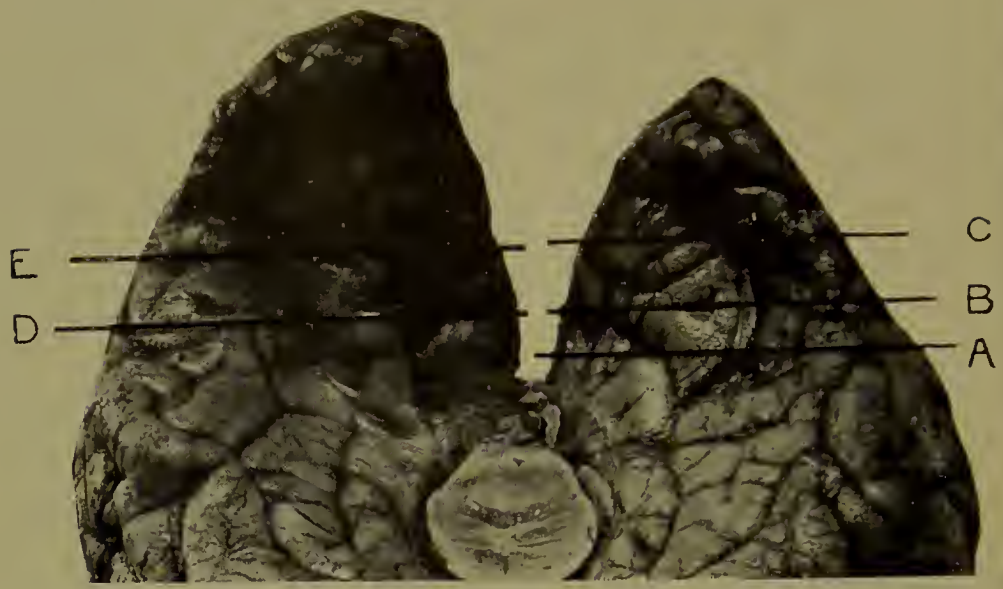

FIG. 3.—UNDER SURFACE OF POSTERIOR PORTION OF CEREBRUM SHOWING SHRINKAGE OF RIGHT OCCIPITAL LOBE AND ATROPHIC CONDITION OF RIGHT TEMPORO-OCCIPITAL CONVOLUTION. LETTERING AND LINES SHOW POSITION OF SECTIONS SUBSEQUENTLY DESCRIBED.

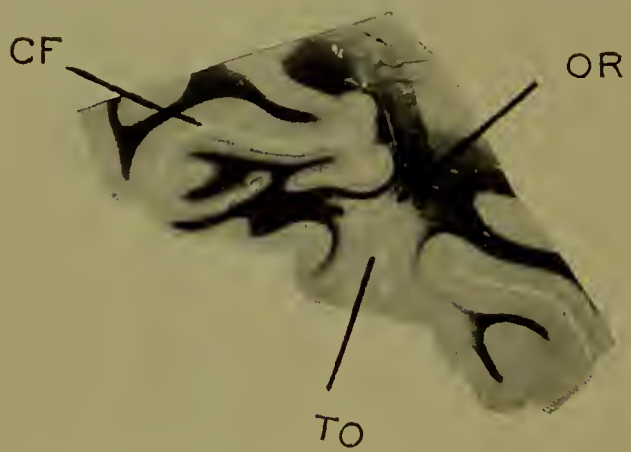

FIG. 4.—SHOWING PORTION OF SECTION A.

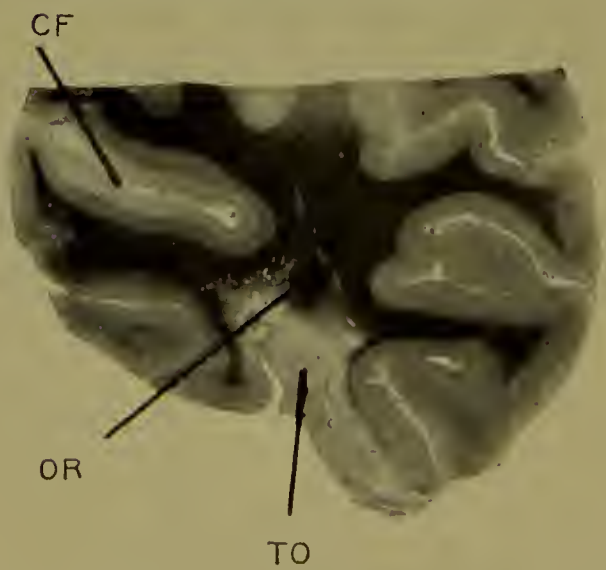

FIG. 5.—SHOWING PORTION OF SECTION B.

PLATE XI.

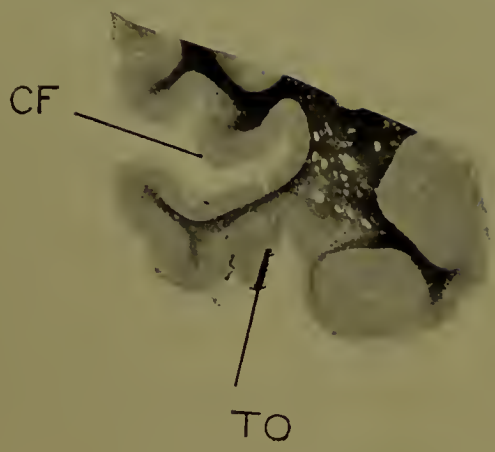

FIG. 6.—SHOWING PORTION OF SECTION C.

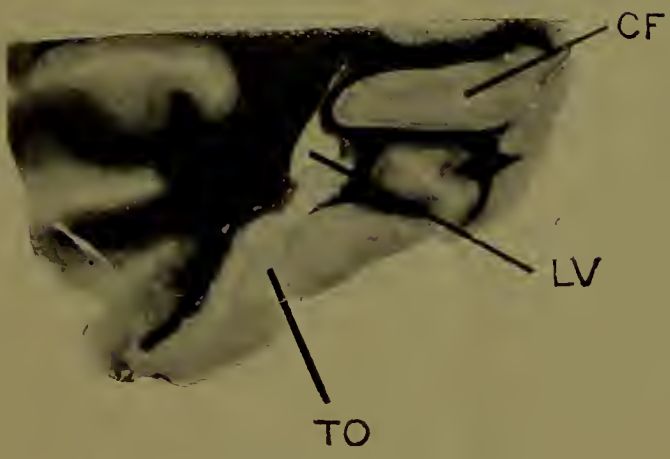

FIG. 7.—SHOWING PORTION OF SECTION D.

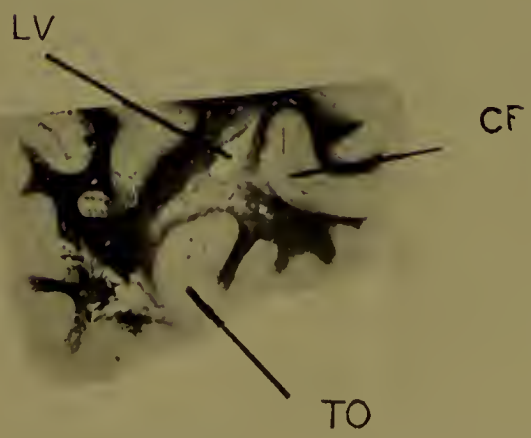

FIG. 8.—SHOWING PORTION OF SECTION E.

Description of Figures:—TO = Temporo-occipital convolution. OR = Optic radiation.
CF = Calcarine fissure. LV = Lateral ventricle.

sight, were found affected by patches of atrophy. The exact relations of these atrophic patches were as follows :—

Right occipital lobe.—Inspection showed that the right occipital was distinctly of smaller size than the left ; this is well shown in fig. 4. It might partly be explained by contortion of the brain in hardening, but the difference between the two sides was so marked that mere contortion is not sufficient for a full explanation, and consequently the difference must be ascribed to either morphological or pathological variation of the lobes. Inspection further showed the existence of a depression on the inferior aspect of the lobe. This depression had a length of $1\frac{3}{4}$ inches and a breadth of $\frac{3}{4}$ inch. The posterior edge of the depression was about $\frac{3}{4}$ inch in front of the posterior extremity of the lobe. The outer edge of the depression reached to within $\frac{1}{2}$ inch from the outer edge of the lobe. In the centre of the depression a crescentic narrow convolution was evident. This atrophied convolution was recognised as being the posterior part of the temporo-occipital or fusiform convolution. To insure accuracy of description and to exclude confusion that might arise from physiological variations of the convolutions, the specimen was submitted to Sir William Turner, F.R.S., who kindly reported as follows :—

"Right hemisphere shows an atrophic condition of the posterior end of a convolution on the tentorial surface. This is bounded internally by the collateral fissure, and externally by the fissure which separates the convolution from the inferior temporo-sphenoidal convolution. The atrophy is strongly marked in that part of this convolution which lies in the same transverse plane as the bottom of the internal parieto-occipital fissure, and which is therefore behind the plane of the posterior end of the gyrus fornicatus. It is not uncommon to find the posterior end of this convolution attenuated in a normally formed brain, but in this brain the change is not a mere attenuation, but is marked by a shrivelled condition of the convolution and an obvious destruction of tissue. The shrivelled appearance extends on the tentorial surface of the occipital lobe to where the convolution is separated from the gyrus lingualis by an intermediate fissure. The convolution is sometimes named in descriptive anatomy the fourth temporal convolution, at other times the temporo-occipital convolution."

To study how far the atrophy extended into the brain surface, three sections were made. One of these (B in fig. 3) was through the centre of the atrophic convolution, another (A) was anterior to that, the third (C) was posterior. These sections were stained both by Heller's method and by alum hæmatoxylin.

Section B, that through the centre of the lesion, showed (*vide* fig. 5) the following points :—

1. A marked atrophy, both of grey and of white matter in a region localised as the temporo-occipital convolution. This convolution was apparently all destroyed with the exception of the association fibres at the inner side of the convolution.

2. No affection of the grey matter of the calcarine fissure.

3. An atrophy of the lower edge of the optic radiation.

Section A, anterior to the one described (*vide* fig. 4), shows the same involvement of brain tissue as seen in section B, namely atrophy of temporo-occipital convolution, no involvement of calcarine fissure, and some involvement of lower edge of optic radiation. It, however, shows that at this level the atrophy of the optic radiation is slightly more extensive than that at the level of section B.

Section C, posterior to B, shows the same atrophy of the occipito-temporal convolution (*vide* fig. 6). It also shows that both the grey and white matter of the calcarine fissure are unaffected, but as the optic radiation is at this level no longer a distinct structure, it does not appear whether it is affected or not.

Left occipital lobe. The examination of the left side of brain showed a lesion of the occipital lobe very similar to that found on the right side, with the exception that on inspection of the surface the atrophy was not so evident, there being no marked depression as on the right side, and no distinctly shrivelled convolution ; it was, however, indicated by some pigmentation. The position of this pigmentation was identified as the posterior end of the temporo-occipital convolution. Sir William Turner kindly corroborated that localisation. Two sections of the left side of the brain were stained and examined, section D (*vide* fig. 7) being made at a plane corresponding as closely as possible to section B of the right side ; the other section, E (*vide* fig. 8), was posterior to D, and intended to correspond with section C of the right side.

Section D showed the following :—

1. A patch of atrophy in the position of the occipito-temporal convolution, extending as far as the floor of the lateral ventricle. The only portion of the temporo-occipital convolution remaining was a small portion, including some of the external association fibres. (*Note.*—Although care was taken to make this section correspond to B of the right side, yet it was found that this section included part of the lateral ventricle, while B did not. This may be ascribed to the lateral ventricle in this brain extending further backwards on the left than on the right side.)

2. That the atrophy, by extending as deep as the lateral ventricle, had destroyed the lower part of the optic radiation, and some of the inferior longitudinal fasciculus.

3. That both the grey and the white matter of the calcarine fissure were unaffected.

Section E showed (1) that the occipito-temporal convolution was partly atrophied, the atrophy being most evident in the association fibres towards the outer side of the convolution.

(2) That the atrophy extended as deep as the lateral ventricle, and extended both on the outer aud inner sides of the ventricle. This implies that part of the optic radiation is involved.

(3) That the white matter between the lateral ventricle and the calcarine fissure was partially destroyed, and the grey matter at the most dependent portion of the fissure for a space of about 3 millimetres was also destroyed, up to and including the line of Vicq d'Azyr.

The brain affection described may be summed up thus : an atrophy of the posterior part of the temporo-occipital convolution of both sides, that on the right side having caused a greater destruction of tissue than that of the left side, but the atrophy of the left side had extended rather deeper than that of the right, having reached the floor of the lateral ventricle, and a small portion of the grey substance of the calcarine fissure. On both sides the lower edge of the optic radiation was affected.

The nature of the atrophy was kindly examined by Dr Ford Robertson of the Scottish Asylums Laboratory, and found to be due to a hyaline degeneration of the adventitia of

the vessels, which had to a great extent cut off the blood supply, and had led to complete destruction of the proper nerve elements, both fibres and cells of the affected portion of the brain.

Remarks.—The case which we here relate may be regarded as one of double homonymous hemianopsia, complete for the colour sense, and incomplete for the form and light senses, in so far that the macular perceptions of light and form were well retained. According to Henschen,[1] a hemianopsia is produced only if there is a lesion of the lower part of the optic radiation or of the calcarine cortex. Our case presents the former condition, but the latter only in trivial degree and apparently only in the left hemisphere. To Henschen also we owe the deduction that the macular region of the retina finds its cortical representation in the anterior part of the calcarine grey matter. Our case does not oppose itself to that conclusion, since, with an almost intact macular perception for form, it exhibits no lesion of the grey matter nor of the immediately subjacent white matter of the anterior portion of the calcarine area.

With Henschen's further assertion that the centres for luminous and for colour perception coincide we would fain agree, for one of us has hitherto seen no cause to alter the opinion expressed in 1888[2] that the existence of a special colour centre was "not proven." But in this particular case the total loss of colour sense is associated with a bilateral lesion of the fusiform convolution so well defined and symmetrical, that it becomes difficult to avoid the conclusion that the grey matter of that convolution is probably concerned in the perception of colours.

The explanation which suggests itself is this. The fibres from the right half of each retina (direct and crossed) constituting the right optic tract find their first destination by way of the lower part of the optic radiation in the right calcarine fissure. Similarly the left optic tract leads first to the left calcarine area. In the grey substance at this part we may with Henschen believe that the "cortical retina" is reproduced, or more precisely that in this centre primary impressions from the half retinæ are formed. It is obvious that

[1] *Rev. Gén. d'Ophth.*, No. 8, 1894.

[2] See *British Medical Journal*, 1888, ii. p. 1036.

to combine these unilateral impressions into a harmonious picture there must be a further conduction of the stimuli along higher paths of association.

Similarly it is quite conceivable that to excite those sensations which we interpret as colour there may be required a conduction of the stimuli out of their first halting-place in the calcarine into the not distant grey matter of the fusiform before the colour value can be appreciated.

Such a theory would accord well with the case before us.

While a great part of the visual portion of the optic radiation has been damaged, the macular fibres have escaped. The eccentric retinal form sense has thus been lost; in the calcarine area central (*i.e.* macular) sense has been retained. But supposing that the fusiform is the seat for the interpretation of chromatic stimuli, the very great destruction which it has undergone in both hemispheres would sufficiently explain the loss of colour-sense by the damage done to *all* the fibres in their second flight from the calcarine to the fusiform.

In this connection it is of the highest interest to note that in Verrey's[1] case of right hemiachromatopsia (the only one recorded with subsequent autopsy) there was found an organised hæmorrhagic cyst $3\frac{1}{2}$ cm. long, 1 cm. broad, and $1\frac{3}{4}$ cm. deep, situated between the floor of the horn of the left lateral ventricle and the basal surface of the occipital lobe. It occupied the white substance of the third occipital convolution; it destroyed more or less completely the white substance of the occipital end of the lingual and fusiform convolutions, as well as that of the postero-inferior part of the cuneus. Towards the base of the brain it approached the median surface of the occipital lobe without actually penetrating the cortex, and there was at parts softening of the white substance at the side of the cyst.

Verrey concluded that "the centre for the colour sense will be found in the most inferior part of the occipital lobe, probably in the posterior part of the lingual and fusiform convolutions."

The lesion which we have described being of still smaller dimensions carries the prediction a stage further.

"One swallow does not make a summer," nor can one

[1] *Arch. d'Ophth.*, 1888, p. 289.

example complete the proof of a theory ; but the facts in this remarkable case, the first, so far as we know, in which a *total* acquired colour-blindness from a cerebral lesion has been supported by pathological examination, point strongly towards the conclusion that if there is a separate centre for colour, its seat is the grey matter of the fusiform convolution.

There is, however, another aspect of the case. The perception of colour is apparently the most delicate portion of the visual act. Interference with colour perception is one of the first symptoms of impaired conduction in the retinæ or optic nerves. Not merely is it the first sense to fail at the onset of distress, but it is the last to return in the process of repair.

It is conceivable, then, that a lesion may readily be of sufficient intensity to damp down the capacity of the cerebral tissue for colour appreciation, without at the same time doing more than slightly lowering the tone for simple luminous and form sensations.

If that be the case, and if the calcarine is the common area for colour as well as simple luminous perceptions, we must seek some other explanation of the function of the fusiform.

When recording this case we take advantage of the opportunity and express our grateful thanks for kind assistance in the examination of the brain lesion to Sir William Turner, Dr Alexander Bruce, and Dr Ford Robertson, each of whom having given us the advantage of their special knowledge of the subjects at issue.